Kidney International (2011) **79,** 140; doi:10.1038/ki.2010.442

Pseudoporphyria associated with hemodialysis

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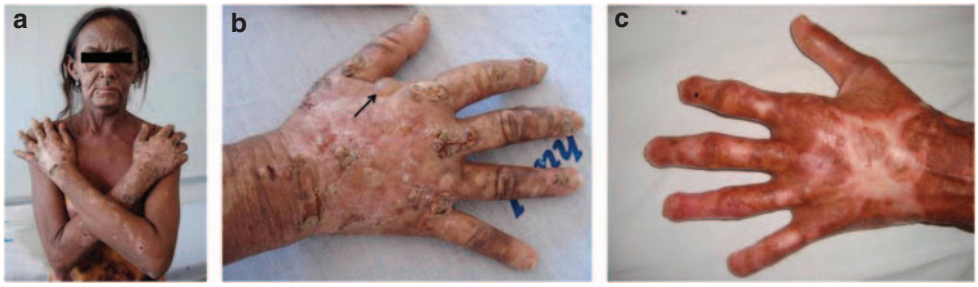

Figure 1 | Pseudoporphyria. (**a**) Hyperpigmentation of the skin. (**b**) Bullae and crusts secondary to scratch lesions. (**c**) Scarring and sclerodermoid appearance after treatment with *N*-acetylcysteine.

A 47-year-old woman with end-stage renal disease due to hypertensive nephropathy, anuric, receiving intermittent hemodialysis for 4 years presented with an itching and burning sensation on sun-exposed sites on her distal limbs and face. This itching and burning was associated with hyperpigmentation of the skin. She reported the sudden onset of multiple bullaes and skin fragility, especially on forearms and dorsal portions of hands (Figure 1a,b). There was no history of use of photosensitizing drugs, and gadolinium, or alcohol abuse. The results of serum tests for human immunodeficiency virus, human T lymphotropic virus, hepatitis C virus, and hepatitis B virus were negative. Porphyrin assays showed normal plasma and fecal levels. Biopsy of lesional skin revealed subepidermal blisters, lymphocytic perivascular infiltrate, and sclerosis of collagen. Direct immunofluorescence was positive showing linear immunoglobulin G deposits in vessel walls and at the basement membrane zone. These results were consistent with a diagnosis of hemodialysis-associated pseudoporphyria, and oral treatment with *N*-acetylcysteine 1200 mg/day was initiated and maintained during 1 year. The skin lesions resolved after 2 months of therapy; however, scarring and sclerodermoids were observed, especially on the fingers (Figure 1c).

Pseudoporphyria is a photodistributed vesicobullous disorder with clinical and histologic features similar to those of porphyria cutanea tarda but without accompanying biochemical porphyrin abnormalities. In the case of suspected pseudoporphyria, a careful medical history, including the amount of ultraviolet light exposure, medications taken, and family history, should be obtained. Physical examination should include an observation of the presence or absence of hyperpigmentation, vesicobullous skin lesions, hypertrichosis, sclerodermoid features, dystrophic calcification, facial scarring, waxy thickening, milia, and skin fragility. Chronic renal failure and dialysis have been implicated in its etiology, but further investigation is necessary to elucidate the pathophysiology fully. Proposed factors include diuretics, aluminum hydroxide, polyvinyl chloride dialysis tubing, hemosiderosis, silicone particles, erythropoietin, and susceptibility to oxygen-free radicals. Treatment includes administration of the glutathione precursor, *N*-acetylcysteine, the discontinuation of contact with suspected agents, and the use of sun protection, especially against UVA wavelengths.